Disease in a dish model provides insight on aging

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Normal aging takes many decades to create major changes in our cells, so it is very difficult to study. As a result, very little is known about this fundamental inevitability of life. But that may change with the help of an unfortunate child, who by the bad luck of a single point mutation developed a rare disease that results in aging at eight to 10 times the normal pace.

A Salk Institute research team lead by Juan-Carlos Izpisua Belmonte has reprogrammed skin cells from the child, who has Hutchinson-Gifford progeria, into induced pluripotent (iPS) stem cells and then forced them to mature into smooth muscle cells in a dish that displayed all the characteristics of aging cells, a model for aging in a dish.

In a Salk press release Belmonte said:

“ Having a human model of accelerated aging may give us new insights into how we age. It may also help prevent or treat heart disease in the general aging population.
In a paper in Nature, the Salk team noted that this progeria is caused by a single point mutation in the gene encoding lamin A, and that there is evidence that defective lamin A also accumulates in the normal aging process via sporadic gene splicing.

The beauty of this model is the researchers were able to provide evidence for the impact of the defective protein. When the reprogrammed cells were in the embryonic-like state the lamin A was silenced, but when those cells were differentiated into smooth muscle the signs of premature aging appeared.

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